TWO CASES OF MUSCULAR ATROPHY OF THE PERONEAL TYPE.

By Given Campbell, M.D.

The rarity with which this type of muscular atrophy occurs, the lack of agreement among neurologists as to its underlying pathological basis, and the advanced age at which my case commences, renders it one of special interest; nor is the interest lessened because of the fact that my patient is a physician, an intelligent and trained observer, able to give a very competent account of the symptoms at a time when the history of such cases is usually incomplete. The following is the history as taken from my case book:

L. C. N., male, aged 45 years, applied for treatment October 5, 1899.

Family history: Patient has several sisters and one brother. All of these are in good health and show no signs of a myopathic heredity. Mother and father healthy. Mother had six brothers and four sisters. These are all healthy. One brother has a daughter that developed epilepsy at puberty. One sister is the mother of nine healthy daughters, all unmarried, and of one son now ten years old. This boy (first cousin to my patient) did not begin to walk until 2½ years old, and at that time it was noticed that his lower legs were wasted and weak. This wasting and weakness have progressed in both legs, being accompanied by fibrillary twitching. This condition was not preceded by pseudo-hypertrophy, but led to distinct talipes varo-equinus, for which an operation was done a few weeks ago. I have not been able to see this boy, but his mother states that he is slightly saddle-backed and that his upper extremities are not weak.

Past history: He never had any serious illness or injury. Five years ago he had a right sciatica with pain and tenderness along the course of the nerve trunk in the thigh. This lasted five days and was not severe. Again twenty months ago, and four months after onset of present disease, he had a second attack of sciatica. This also was located in the right limb and caused pain and tenderness along the nerve trunk in the thigh, and had a mild course and duration of five days. He has had no acute infection for the last twenty years, and has never had syphilis. He was a moderate drinker for ten years prior to onset of present malady, but has drunk nothing alcoholic since. During this time he averaged one glass of whiskey and two glasses of beer daily, though he never went on sprees. He was unusually susceptible to effects of alcohol. He was never exposed to the action of lead and was moderate in the use of tobacco.

Symptoms: Up to September, 1897, he was well, and led the active life of a country practitioner. At this time, however, he noticed that he turned his ankles very easily, that his thigh muscles showed an unusual tendency to cramp, and that he was not able to lift his toe properly in walking. He began to wear out the toe of his shoe. He noticed a wasting of his anterior tibial muscles and tested his knee-jerk and found it absent. He is sure, however, that there was no considerable weakness in the extensors of the knee at this time. Thinking that he had tabes, he tested himself carefully for the presence of any sensory symptoms, but failed to find any. His sensibility to touch, pain and temperature was normal, and he had no ataxia. He was able to stand on one foot with his eyes closed. A few weeks later these tests were all verified by a brother physician, and again no sensory symptoms were found, but the knee-jerk was still absent even with reinforcement. The weakness and wasting continued to increase in the legs symmetrically and was gradually progressive. By the end of five or six months the anterior tibial group was almost as atrophic as at present, and the muscles of the anterior aspect of the thigh were beginning to be affected. About this time the patient noticed twitchings in the gluteal muscles and the lumbar group. These twitchings have persisted ever since, and are severe enough to be troublesome to him. They occur during sleep, and have been present in the shoulder girdle muscles for the last year. They are most severe in the right arm (biceps and deltoid). This twitching is worse when he has been agitated or is a little run down. The disease has gradually progressed in the lower limbs so that they are now quite useless, and he can barely move the toes in flexion. No other movement is possible. He states that his sexual functions have been normal up to one month ago, but that since then there has been a slight falling off in venereal desire, and at present date, December 5, this decrease has progressed to a complete loss of all desire. Sphincters are normal. He has had no sensory involvement of any kind at any time in the course of the disease, although this was carefully and frequently searched for by an observer expecting to find it.

Physical examination: He is of healthy aspect and fairly well nourished. His face shows no signs of muscular atrophy, his eyes are normal, he has no nystagmus, and his pupillary reflex is normal. An examination of the fundus was made by Dr. Wolfner and revealed nothing abnormal. Vision is normal. No signs of any cranial nerve lesion are present.

Upper extremities: No distinct signs of muscular atrophy are present, but both deltoids seem shorter than normal. The small muscles of hand are neither atrophic nor weak. Grasp: right hand 115, left 93. The left deltoid is distinctly weak. He can hold his right arm out from the shoulder, supporting a weight of seven and three-quarter pounds. Considering that he uses his arms a great deal in wheeling himself about in an invalid chair, he is not strong, even in the right deltoid. With the left arm he can support under similar conditions only two and seven-eighths pounds. The left supinator longus is only about one-third as strong as the corresponding muscle on the right side. It is interesting to note that the twitchings, which are distinctly visible now and are of a coarse fibrillary nature, are chiefly located in the right arm, the one less paralyzed. Body muscles: Thoracic muscles are normal; the lumbar group is not atrophic; the abdominal muscles seem perhaps somewhat atrophic. Lower limbs: The atrophy is marked in both lower extremities. Perhaps the adductors are rather less atrophic than the others. The lower leg contains very little if any muscular tissue. The limbs are perfectly helpless.

Reflexes: The deep reflexes are everywhere absent. Cremasteric reflex is present.

Electrical reaction: Some twitching is seen in the quadriceps muscles of the lower limbs, but these muscles fail to react to either galvanism or faradism. In the upper limbs, including left deltoid and supinator, the reactions to galvanism and faradism are normal. No reaction of degeneration and no sensory impairment can be found. Some shortening of calf muscles is present, so that the foot cannot be brought quite to a right angle with the bone of lower leg. Attempts to bend it farther cause pain referred to the belly of calf muscles.

The pulse now varies between 85 and 100, but the patient states that it was always frequent. The viscera are normal and the urine shows nothing abnormal. He has no tenderness along the nerve trunks.

The most interesting points in regard to this case are: 1st. The advanced age at which the disease began, forty-three years. I can find no record of a case beginning this late. 2d. The rather rapid progress of the disease. 3d. The distinct weakness of the left deltoid and the supinator of that side, muscles not usually affected in this disease, and the occurrence of this weakness before there is any perceptible atrophy of the hand or forearm. 4th. The complete absence of sensory symptoms. 5th. The abolishment of the knee-jerk at a time when the extensors of the knee were apparently in perfect condition and when no sensory disturbance was present. The fibrillary twitching was permanent and quite constant; it was distinctly visible, and was perhaps the symptom pointing most strongly to an involvement of the spinal cord. Were it not for this and the tendency to cramps of the muscles the absence of all sensory impairment might make this case seem one in which the muscles were primarily involved. The question naturally arises as to why the deep reflexes were absent when the voluntary muscular power was good. The fact that no sensory disturbance has ever occurred would seem to imply that the defect was not on the afferent side of the reflex arc, notwithstanding the autopsy findings by Marinesco and by Sainton, in whose cases a degeneration of the posterior columns with changes in the cells of the anterior horns was found. But in Marinesco's case a distinct sensory involvement was present. In Sainton's case it was not excluded. In investigating the efferent side of the arc, we find a good condition of psychomotor impulses, and this would imply that the muscle, motor plate and efferent neuron were normal up to their connections with the pyramidal tract.

The writer is well aware that Paul Sainton[1] has attempted to exclude from the category of Charcot-Marie amyotrophy cases in which the hands are not atrophied and in which the shoulder girdle muscles are involved. It is also plain that in my case the progress of the disease has been more rapid than is perhaps the rule, and that the atrophy does not follow the garter type in the lower limbs to which Charcot first called attention. The clinical picture is one of weakness and wasting going hand in hand, perfectly symmetrical, and of insidious progress, commencing in the peroneal group of muscles and extending upward, accompanied by no sensory impairment, with early loss of knee-jerk, complicated by distinct fibrillary twitchings and by the tendency of the muscles to cramp. This picture, taken in connection with the fact of a similar malady occurring in a first cousin, leaves room for no other diagnosis than that of a muscular atrophy of the peroneal type, to which the French have given the name of its discoverers, Charcot and Marie, and the English that of its principal British exponent, Howard Tooth. I might remark that Tooth[2] has reported several cases in which the upper extremities were not involved. Sachs'[3] case was one in which there was no wasting of the hands, but there was wasting of the entire thighs and some slight wasting of the infraspinatus muscles. In Jacoby's[4] case the upper extremities were not affected, and some lordosis was present.

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[1] Thèse du Doctorat, 1899.

If the consideration of this case brings out any especial points of interest they are:

1st. That muscular atrophy of the peroneal type may commence so late in life as forty-three years.

2d. That the atrophy is not always of the garter type in the lower limbs (extending a little above the knee), but may involve the entire thigh.

3d. That well-marked cases may occur in which the hands are not atrophic.

4th. That the knee-jerks may be early abolished in cases which are, and subsequently remain, free from all sensory impairment.

Since reporting the above a second case of this disease has been referred to me for treatment. My two patients are in no way related to each other and live in different parts of the country. The clinical history of this second patient is as follows:

W. K., male, aged 32, height 5 feet 10 inches. Family history: Mother's sister epileptic. Otherwise family history is unimportant. Past history: Patient was born at term after normal gestation and labor. Well until 2 years of age. Nursed on breast by mother until 1 year old, but mother's milk was deficient in quantity and deficiency was made up by oatmeal water, this being given from time of birth. At 2 years of age patient had an attack of epidemic meningitis, one month's duration, good recovery, no sequelæ. Patient was a perfectly healthy and active boy until 15 years of age, at which time his mother noticed that his instep was gradually becoming higher. She had to have shoes made to order for him. This deformity has steadily progressed, the feet becoming smaller and the instep more arched. Progress gradual and steady. For several years after onset of the atrophy there was very little inconvenience in walking. This has increased lately, and is due rather to the deformity than to any muscular weakness. Six years ago patient began to be much troubled by cramps, chiefly in the muscles of the legs. This tendency, while not so severe, is still distinctly present, and can be elicited at will by patient forcibly contracting flexors of knee or calf muscles. During the last four years distinct fibrillary twitchings have been present, principally in the muscles of the arms.

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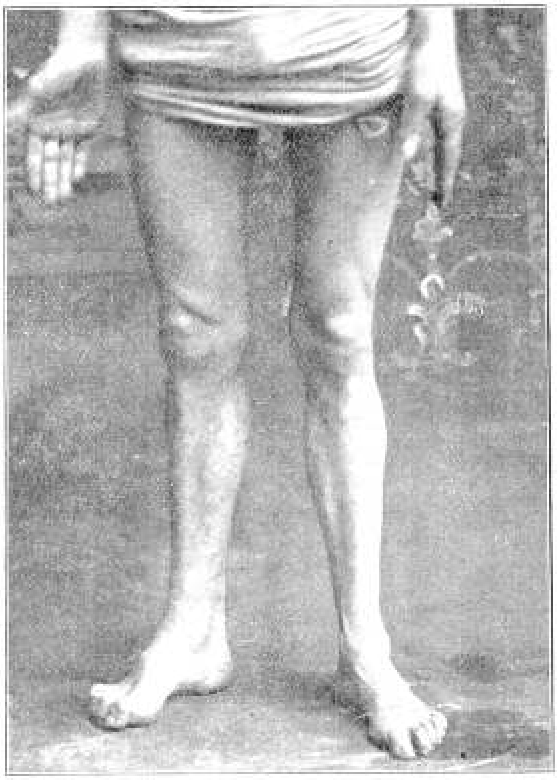
[2] Thesis Cambridge, 1880.

[3] Brain, Jan., 1890.

[4] JOURNAL OF NERVOUS AND MENTAL DISEASE, 1894.



Case II.—The deformity of claw-foot is especially well shown on the right side in the photograph.

There has never been any pain or other sensory involvement. Physical health has remained good. For the last fifteen years he has been gradually growing deaf, and this loss of hearing has been complete for the past three years. Several years ago he noticed a dimness of vision, for which he consulted an oculist, by whom a diagnosis of double stellate capsular cataract was made. Vision is now very imperfect. Rectal and vesical symptoms are absent. Sexual power is normal. Patient is moderate in the use of tobacco and does not use alcohol, tea or coffee.

Physical examination reveals no disease of the vital organs. Urinary secretion normal. The muscle reflexes are very greatly diminished. Knee-jerks barely obtainable by reinforcement. The superficial reflexes, however, are distinctly increased, tickling the sole of the foot almost producing a clonic spasm of the flexors of knee. Sensation is normal to touch, pain and temperature. No tenderness of the nerve trunks is present. Measurements: Calf of leg, right, 31.5 cm., left, 31.5 cm.; thigh 5 cm. above upper margin of patella, right, 32 cm., left, 31 cm.; thigh 7.5 cm. below gluteal fold, right, 46 cm., left, 46 cm. The neck measures 34.5 cm. Electrical reactions normal. There is a slight but perceptible wasting of the small muscles of the hands. The small muscles of the feet are very atrophic, and the deformity of claw-foot is very distinct and pronounced. The small size of the lower leg is probably due to disuse rather than to the direct effects of the disease, for it will be noticed that the entire limb, excepting the foot, is symmetrically small.

A comparison of this case with the one first reported will be of interest in considering the rapidity of onset in this disease. Both cases are extreme, the first being unusually rapid in its march and the second very slow; for after a period of seventeen years the atrophy has not extended noticeably beyond the small muscles of the feet and hands. The existence of cataract and deafness is probably a mere coincidence, although the fact that the deafness is total and involves bony conduction makes it probable that it is of central origin.